POSTERIOR SEMICIRCULAR CANAL AND VESTIBULAR AQUEDUCT DEHISCENCES CAUSED BY JUGULAR BULB ABNORMALITIES

UŽPAKALINIO PUSRATINIO KANALO IR PRIEANGIO VANDENTIEKIO DEHISCENCIJOS DĖL JUNGO VENOS STORMENS ANOMALIJŲ

Irina Arechvo

Karoliniškių poliklinikos Otorinolaringologijos konsultacijų skyrius
Outpatient Otorhinolaryngology Department, Karoliniskiu Polyclinic

SANTRAUKA

Reikšminiai žodžiai: vidinės ausies dehiscencija, jungo venos stormens anomalijos, vertigo, pulsuojantis ūžesys.

Straipsnyje pristatomi užpakalinio pusratinio kanalo ir prieangio vandentiekio dehiscencijų atvejai dėl jungo venos stormens anomalijų. Taip vadinamos trečiojo lango ligos yra neseniai aprašytos patologinės būklės, sukeltos dalinio kaulinio labirinto ar sraigės kapsulės nebuvimu. Šio straipsnio tikslas – pateikti jungo venos stormens anomalijų sukeltų dehiscencijų simptomus ir jų vystymosi eigą. Taip pat dėl galių užpakalinių pusratinių kanalų ir prieangio vandentiekio dehiscencijų, sukeltų jungo venos stormens anomalijų, buvo peržiūrėti aukštos rezoliucijos kompiuterinės tomografinės vaizdai pacientų, sergančių lėtiniu vidurinės ausies uždegimu su arba be cholesteatomo. Retrospektyviai buvo ieškoma koreliacijų tarp šių neurovizualinių vaizdų ir otoneurologinių simptomų.


ABSTRACT

Key words: inner ear dehiscence, jugular bulb abnormalities, vertigo, pulsatile tinnitus.

The study presents a case series of posterior semicircular canal and vestibular aqueduct dehiscences caused by jugular bulb abnormalities. The so-called third window lesions are recently described pathological conditions caused by the partial absence of the bone of the labyrinth or cochlear otic capsule.

The objective of the study was to present symptoms and their chronological development in patients with jugular bulb abnormalities. Also the high-resolution computed tomography scans performed in patients with chronic otitis media with or without cholesteatoma were reviewed for possible posterior semicircular canal and vestibular aqueduct dehiscences. Radiological findings were retrospectively correlated with the otoneurological symptoms in these patients.

Three patients were symptomatic and four – without any specific complaints. The high-riding jugular bulb, the jugular bulb diverticulum were often radiological findings. In some patients they might cause otoneurological symptoms. The clinical, audiological and imaging characteristics of the inner ear dehiscences are critically discussed. The importance of the accurate correlation between the symptoms and radiological findings in each dehiscence case is also emphasized.

INTRODUCTION

During the last two decades a growing number of the reports on the superior semicircular canal dehiscence appeared in the scientific literature [1–3]. However, an amount of the posterior semicircular canal dehiscence (PSCD) case reports remains small [4–7]. To date, as far as I know, fewer than 120 ears with PSCD can be found in the English-
Most of them are related to the jugular bulb abnormalities – a high-riding jugular bulb and jugular bulb diverticulum. The clinical findings definitely related to the inner ear dehiscences remain controversial [6].

The objective of the study was to present symptoms and their chronological development in the patients with jugular bulb abnormalities. Also I reviewed the high-resolution computed tomography (HRCT) scans performed in patients with chronic otitis media with or without cholesteatoma for possible posterior semicircular canal and vestibular aqueduct dehiscences. Radiological findings were retrospectively correlated with the otoneurological symptoms in these patients.

CASE REPORT 1

A 48-year-old woman was referred to the otorhinolaryngologist with the symptoms of hearing loss, roaring tinnitus, aural fullness in her right ear, and vertigo attacks that lasted from several minutes till several hours and were position-dependent. Vertigo was provoked by the oblique head movements backward at the right side. She denied autophonia and hearing of chewing or ocular movements. For the first time the disease manifested ten years before with the acute positional vertigo that was caused by sudden tilting of the head back and/or forward. In one year aural fullness and hearing loss suddenly occurred. She was diagnosed with sudden sensorineural hearing loss. She was treated by vasodilators and anxiolytics with partial resolution of the audiological symptoms. However, she started to feel low-frequent pulsatile tinnitus in her right ear especially during the vertigo attack. She was treated with Betahistine and Nicergoline. She also observed that the loud external sounds induced dizziness.

The pure-tone audiogram revealed moderate-to-severe sensorineural hearing loss up to 35 dB without air-bone gap in the low-frequency range. Videonystagmography and caloric testing showed bilateral vestibular weakness. Tullio phenomenon was positive on the right side. Other otoneurological tests were normal. Magnetic resonance imaging (MRI), that was carried out to exclude cerebellopontine angle pathology, revealed a large diverticulum of the high-riding medial jugular bulb on the right side that was in contact with membranous posterior semicircular canal and vestibular aqueduct (Fig. 1).

The imaging findings were thoroughly discussed with the patient and the periodical follow-ups were planned. She reported improvement in her equilibrium after vestibular rehabilitation had been started.

Fig. 1. 3D volume rendering technique (A) showing the diverticulum on the right side (arrow) and normal jugular dome on the left. High jugular bulb (asteriks), contacting right membranous posterior semicircular canal, T2 3D CISS MR image (B). A contact point (arrow) between the membranous endolymphatic duct/sac and jugular bulb diverticulum, T1 weighted MRI image (C)

CASE REPORT 2

A 41-year-old man presented to the otorhinolaryngologist with a 2-year history of hearing loss, pulsating tinnitus on the right side, vertigo and disequilibrium. At the beginning hearing loss presented as sudden then became fluctuating, and finally – constant.

Microscopically, the external auditory canals and middle ears were without any pathologic changes. Tympanometry revealed A-type curve at both sides. The Hennebert’s sign was positive at the right side. The pressure changes in the external auditory canal caused horizontal eye movements to the left with torsional component. The stapedial reflex was absent at the right side. Flat mixed hearing loss up to 61.7 dB with an air-bone gap of 16.7 dB at the low frequencies was identified using standard audiometry testing. A preliminary diagnosis of cochlear otosclerosis and/or cerebellopontine angle pathology has been proposed. However,
MRI revealed a high jugular bulb (HJB) on the right side that was dehiscent to the posterior semicircular canal (Fig. 2).

The patient refused further evaluation by HRCT. The disease was discussed with the patient and hearing aid was fitted on the right ear.

CASE REPORT 3
A 42-year-old woman has been consulted by an otorhinolaryngologist due to the sudden vertigo attack with coordination impairment. On physical examination spontaneous horizontal nystagmus to the right side with positive head thrust and stepping tests on the left have been determined. Vestibular aqueduct dehiscence due to the HJB on the left has been shown on the HRCT scans (Fig. 3). She improved significantly over next 12 hours with complete resolution of the symptoms within one week.

ASYMPTOMATIC CASES
I also retrospectively reviewed the HRCT of the temporal bones of 91 patients consulted due to the chronic otitis media with or without cholesteatoma. I found 18 (19.8 \%) cases of the HJB. In the present report HJB was considered to be as above the level of the basal turn of the cochlea, superior tympanic annulus and/or reaching the level of the internal auditory canal (IAC) [4, 6, 8]. Among them four cases (22.2 \%) of non-iatrogenic and non-traumatic vestibular aqueduct dehiscence (VAD) were determined (Fig. 4). An absence of intervening bone was observed in at least 2 consecutive images and in multiple planes, the tactic proposed by previous researchers [6]. One patient possessed bilateral VAD combined with a posterior-inferior erosion of the internal auditory canal by HJB (Fig. 5). This patient complained autophony and ear fullness. The latter symptoms were attributed to the patulous Eustachian tube that was confirmed endoscopically and microscopically. None of patients complained vertigo, pulsatile tinnitus or other otoneurological symptoms confident with VAD and/or Ménière’s syndrome.

DISCUSSION
Previous researchers reported that PSCD as well as VAD due to the jugular bulb abnormalities might be symptomatic as well as completely silent (Table 1). My experience confirmed this data. In the present report four patients were asymptomatic or had unrelated complaints, whereas three possessed the symptoms of Ménière’s syndrome, and/or inner ear dehiscence. Therefore, I emphasized that every radiologically evident posterior semicircular canal or vestibular aqueduct dehiscence should be thoroughly correlated with the clinical presentation, as the most patients are asymptomatic.

The progression of the symptoms in the first case could be explained by progression of the jugular bulb diverticulum
since the previous investigators suggested non-anatomically stable and progressively modeled by centrifugal hemodynamic forces medial jugular bulb [14]. In this patient the diverticulum circumvented the membranous labyrinth inferiorly what is seen on the MR image (Fig. 1B). Also the high diverticulum came into the contact with the distal part of the endolymphatic duct and sac. The latter finding explained the symptoms of the Ménière’s syndrome. Pulsatile tinnitus could be explained by a transmission of the venous pulsation and/or turbulent flow to the inner ear structures [10]. As it was proposed earlier the effect on the endolymphatic duct/sac may be direct or indirect, causing a decrease of the endolymph secretion [14]. In the present case a direct connection between jugular bulb diverticulum and endolymphatic sac was detected (Fig. 1C). The authors of the same study proposed possible relation between high medial jugular diverticulum and sensorineural hearing loss with vertigo even in the case of absence of the exact evidence of the dehiscence.

The third window lesions can be misdiagnosed with Ménière’s disease, perilymphatic fistula, patulous Eustachian tube, ossicular fixation, and more often otosclerosis [5]. The history of the first patient was also suspicious for benign paroxysmal positional vertigo diagnosis. However, a Dix–Hallpike symptom was negative bilaterally in this case.

In the second case stapedial reflex was absent what was in accordance with the otosclerosis diagnosis. In recent histopathological study Friedmann et al. showed that otosclerosis was not more common in patients with HJB [6]. The absence of the stapedial reflex is a sign of otosclerosis, which may coexist with jugular bulb abnormalities. Recently, Van Rompaey et al. reported a patient with HJB dehiscent to the vestibular aqueduct and confirmed otosclerosis. The stapedotomy in this patient was unsuccessful [15]. The authors emphasized an importance of HRCT before considering revision stapes surgery. This is an essential imaging technique as well to confirm the diagnosis of the dehiscence. Recently Browaeys et al. proposed MRI as screening tool for dehiscence exclusion (sensitivity of 100 % and specificity of 99.1 %) [16]. Although MRI can give a false positive result for the inner ear dehiscence, in the present cases the

Table 1. Posterior semicircular canal and vestibular aqueduct dehiscence cases due to the jugular bulb abnormalities reported in English-language literature

<table>
<thead>
<tr>
<th>Authors, year</th>
<th>Number of patients with inner ear dehiscences</th>
<th>Number of symptomatic patients</th>
<th>Type of study</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wadin et al., 1986 [4]</td>
<td>PSCD – 4; VAD – 12</td>
<td>PSCD – 1 (25 %) with hearing loss, unsteadiness and fistula symptoms; VAD – 12 (100 %) with Ménière’s disease</td>
<td>Clinical and radiologic</td>
<td>?</td>
</tr>
<tr>
<td>Hourani et al., 2005 [8]</td>
<td>VAD – 23; among them 1 – bilateral dehiscence</td>
<td>9 (39.1 %) – dizziness; 11 (47.8 %) – hearing loss, with 9 (81.8 %) of these 11 – hearing loss on the same side as the dehiscence (7 – SNHL; 2 – CHL); 1 (4.3 %) – tinnitus on the same side and 2 (8.69 %) – ear fullness</td>
<td>Clinical and radiologic</td>
<td>?</td>
</tr>
<tr>
<td>Crovetto et al., 2010 [9]</td>
<td>PSCD – 1</td>
<td>?</td>
<td>Radiologic</td>
<td>?</td>
</tr>
<tr>
<td>Friedmann et al., 2010 [10]</td>
<td>PSCD – 4; VAD – 9</td>
<td>All (100 %)</td>
<td>Clinical and radiologic</td>
<td>?</td>
</tr>
<tr>
<td>Gopen et al., 2010 [5]</td>
<td>PSCD – 7</td>
<td>All (100 %)</td>
<td>Clinical and radiologic</td>
<td>?</td>
</tr>
<tr>
<td>Nomiya et al., 2010 [11]</td>
<td>PSCD – 1</td>
<td>Asymptomatic</td>
<td>Histopathologic</td>
<td>?</td>
</tr>
<tr>
<td>Friedmann et al., 2012 [6]</td>
<td>PSCD – 4; VAD – 25; FND – 5</td>
<td>5 (50 %)</td>
<td>Clinical and radiologic</td>
<td>?</td>
</tr>
<tr>
<td>Lim et al., 2012 [12]</td>
<td>PSCD – 1</td>
<td>1 (100 %)</td>
<td>Clinical and radiologic</td>
<td>Surgical</td>
</tr>
<tr>
<td>Sone et al., 2012 [7]</td>
<td>3 (pediatric): both PSCD and VAD – 2; VAD – 1</td>
<td>All (100 %)</td>
<td>Clinical and radiologic</td>
<td>?</td>
</tr>
<tr>
<td>Gubbels et al., 2013 [13]</td>
<td>PSCD – 1</td>
<td>1 (100 %)</td>
<td>Clinical and radiologic</td>
<td>Surgical</td>
</tr>
</tbody>
</table>

? – Information is unknown; SNHL – sensorineural hearing loss; CHL – conductive hearing loss; FND – facial nerve dehiscence
MRI findings were correlated well with the clinical data. In two symptomatic patients the dehiscences were clearly seen on MR images (Figs. 1B and 2B, 2C). In the present report 3D volume rendering technique clearly showed the contact place between the membranous labyrinth and jugular bulb (Fig. 2D) and correlated well with the conventional 2D techniques. MRI T2 consequences did not show any disturbances in the labyrinthine fluid signal, therefore the posterior semicircular canals and endolymphatic duct and sac fluid spaces were free and non-obligated.

In the present report two symptomatic HJB were observed on the right. Moreover, in the second case the MRI showed hypoplastic venous sinus system on the left side. These findings correspond with the previous data that the venous sinuses and the jugular vein are larger on the right side in 60.5–75 % of individuals [8]. PSCD and VAD are also more often diagnosed at the right side as well [5, 6].

In the present report the asymptomatic HJB cases and even dehiscence to the vestibular aqueduct were interpreted as incidental finding. I emphasize that in every case radiological data should be thoroughly correlated with the clinical symptoms. Moreover, in recent histologic study Friedman et al. showed that dehiscent vestibular aqueduct is well tolerated and endolymphatic hydrops symptoms are quite rare in such patients [6]. In the same study the authors showed that posterior semicircular canal dehiscence was the most frequently symptomatic finding. This correlates with the results of the present report. Most VAD were asymptomatic whereas if they were combined with PSCD the patients reported otoneurological symptoms.

Although, successful results with the complete resolution of the symptoms after the surgical resurfacing of the eroded posterior semicircular canal have been reported previously [12, 13, 17], a conservative tactic has been chosen in the current study.

ACKNOWLEDGMENT

This manuscript is dedicated to the memory of great otologic researcher and ear surgeon Saumil N. Merchant. His brilliant experimental and clinical works inspired the physicians all around the world.

REFERENCES